Br Heart J 1987;57:364–70

The dysplastic pulmonary valve: echocardiographic features and results of balloon dilatation

NORMAN N MUSEWE, MURRAY A ROBERTSON, LEE N BENSON, JEFFREY F SMALLHORN, PATRICIA E BURROWS,* ROBERT M FREEDOM, C A F MOES,* RICHARD D ROWE

From the Division of Cardiology, Variety Club Cardiac Catheterization Laboratories, Hospital for Sick Children, Toronto; and the Department of Paediatrics, University of Toronto Faculty of Medicine, Ontario, Canada

SUMMARY The feasibility of using balloon dilatation to relieve stenosis caused by dysplasia of the pulmonary valve was assessed in seven patients (five female, mean age two years) with angiographically confirmed dysplasia who were identified among 38 patients with pulmonary valve stenosis selected for balloon dilatation over a two year period. The clinical features in three patients were consistent with Noonan's syndrome. In all patients the gradient across the valve was assessed by cross sectional echocardiography and Doppler echocardiography before cardiac catheterisation. Balloon dilatation was performed by conventional techniques. In one patient, who had balloon dilatation in the operating room before surgical valvectomy, the diameter of the valve orifice increased from 3 mm to 10 mm. Inspection showed a tear along the anterior commissure. The mean (SD) pressure gradients between the right ventricle and pulmonary artery before and immediately after dilatation in five patients were not significantly different (58 (28) and 47 (12) mm Hg) respectively. There was no overall significant change in the degree of stenosis when four of these patients were examined by Doppler echocardiography six months after operation (44 (17) mm Hg), although one patient (case 5) did show a significant reduction in gradient. This patient had angiographic and echocardiographic features of dysplasia and commissural fusion. Several echographic features were common to all patients and distinguished them from cases of typical pulmonary valve stenosis. These were: *(a)* pronounced thickening of leaflets; *(b)* leaflet immobility in diastole and systole; *(c)* no dilatation of the sinuses of Valsalva in diastole, and *(d)* supra-annular narrowing.

These poor results of balloon dilatation suggest that commissural fusion is not an important mechanism for causing stenosis in the dysplastic pulmonary valve. When dysplasia of the pulmonary valve is identified clinically and echocardiographically, balloon dilatation is unlikely to improve haemodynamic function; however, it should be attempted if commissural fusion is present.

About 10% of cases of congenital heart disease have isolated pulmonary valve stenosis.[1] The mechanism of stenosis may be commissural fusion, resulting in a reduced haemodynamic orifice, or dysplasia of valve leaflets,[2-6] which presents obstruction to flow, with or without a commissural component and a hypoplastic valve annulus. Dysplasia of the pulmonary valve is often found as an associated cardiac anomaly in Noonan's syndrome.[7 8] The angiographic features of dysplasia include pronounced thickening of valve leaflets with irregular asymmetric doming in systole, little change in leaflet appearance between diastole and systole, hypoplasia of the proximal main pulmonary artery, and no radiolucent jet on cineangiocardiography.[5] Surgical experience with standard commissurotomy had been disappointing[6 9-11] and this technique has been abandoned in favour of partial or complete valve tissue resection, with or without patch enlargement of the right ventricular outflow tract.

Requests for reprints to Dr Lee N Benson, Division of Cardiology, Hospital for Sick Children, 555 University Avenue, Toronto, Ontario M5G 1X8, Canada.

*Department of Radiology, The Hospital for Sick Children, Toronto and Department of Radiology, University of Toronto Faculty of Medicine.

Accepted for publication 11 November 1986

Table 1 *Results of balloon dilatation in seven patients with dysplastic pulmonary valves*

		Pre-dilatation		*Post-dilatation*		
Patient No	*Annulus size (mm)*	*RV pressure as % of systemic*	*Gradient (mm Hg)*	*RV pressure as % of systemic*	*Gradient (mm Hg)*	*Follow up gradient (mm Hg)*
1	15	90	60	—	—	—
2	10	69	36	67	48	42
3	11	60	40	50	32	47
4	11	142	150	—	—	—
5	10·4	103	96	70	60	23
6	8	86	80	72	60	65
7	10	60	38	60	38	—
Mean (SD)	10·7 (2·11)	87 (29)	71·4 (41·5) 58 (28)†	63·8 (8·9)	47·6 (12·6)	44·2 (17·2)

RV, right ventricular.
†Only patients dilated.

Percutaneous balloon dilatation has now replaced open heart valvotomy for the relief of typical pulmonary valve stenosis.[12-17] The attendant morbidity is minimal and short term results appear to be good. A few patients with dysplasia of the pulmonary valve have undergone this procedure but balloon dilatation has not been specifically assessed in this condition.[14 15 17] We report our experience of using balloon dilatation in patients who had dysplasia of the pulmonary valve that was identified by cross sectional echocardiographic criteria.

Patients and methods

Seven of 38 patients undergoing percutaneous balloon dilatation of the pulmonary valve between January 1984 and December 1985 were prospectively identified as having clinical features consistent with dysplasia of the pulmonary valve. Three patients had the Noonan phenotype (cases 1, 2, and 3). None had a pulmonary ejection click. Plain chest radiographs did not reveal post-stenotic dilatation of the main pulmonary artery (a feature noted in at least 90% of patients with typical pulmonary valve stenosis[5]). The frontal QRS axis of the electrocardiogram exceeded +170° in five cases. Two patients (cases 3 and 5) had an associated patent foramen ovale.

Echocardiographic imaging was carried out with an ATL Mark 600 or Ultramark 8 mechanical sector scanner ultrasound system (Advanced Technology Laboratories Inc, Bellevue, WA). Images were obtained from the standard and high parasternal short axis views. When we simultaneously measured Doppler echocardiographic gradients and intracardiac pressures in 39 patients with pulmonary valve stenosis we obtained a good correlation ($r = 0·95$) (gradient (catheter) = 0·89 [gradient (echo)] + 3·98).

Angiocardiography was carried out under ketamine anaesthesia before balloon dilatation. The details of our dilatation procedure have been reported elsewhere.[18] Briefly, systemic and right ventricular pressures are continuously monitored before and during dilatation. Balloon catheters with diameters that are 20–30% greater than annulus diameter measured from the lateral right ventriculogram (table 1) are positioned across the pulmonary valve by means of a guide wire in the left pulmonary artery. Inflation-deflation cycles are rapid (less than 15 seconds) and they are repeated (usually for two or three cycles) until no waist can be identified when the balloon is inflated.

Patient 1, in whom pulmonary valve dysplasia had been identified clinically and angiocardiographically, underwent balloon dilatation at the time of surgery. Balloon size was selected according to the preoperative angiogram and the balloon was placed across the pulmonary valve ring from the pulmonary artery and inflated once after cardiopulmonary bypass had been started.

Results

Table 1 shows the results of balloon dilatation. The balloon catheter could not be passed across the pulmonary valve in one patient (case 4). In the operating room, before dilatation, patient 1 was seen to have thickened "fleshy" valve leaflets. A tear was seen in the anterior commissure of this valve as a result of dilatation. This suggests an element of commissural fusion, and in this instance the orifice increased from 3 to 10 mm according to Hegar dilator sizing.

During percutaneous balloon dilatation in the remaining five cases no waist was seen during the initial inflation cycle. Immediately after the procedure the mean pressure gradient across the pulmonary valve decreased insignificantly from 58 (28) to 47 (12) mm Hg. Doppler echocardiography in four patients (cases 2, 3, 5, and 7) six months after operation showed no overall significant relief of stenosis. One

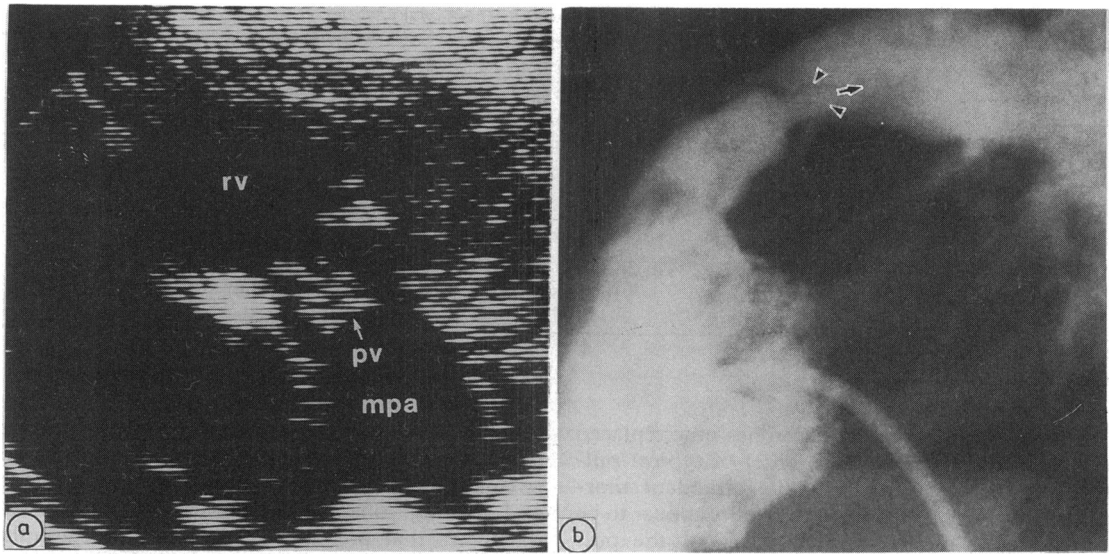

Fig 1 *Cross sectional echocardiograms. (a) Precordial short axis view of a valve showing dysplasia (white arrow) and commissural fusion. The cusp closest to the aorta appeared to be immobile during systole. The left sided cusp, however, appeared to dome during the cardiac cycle. (b) An angiocardiogram from the same patient, showing a central jet (black arrows); the valve leaflets are thickened. mpa, main pulmonary artery; pv, pulmonary valve; rv, right ventricle.*

patient (case 5), however, did have a significant reduction in gradient from 60 mm Hg immediately after dilatation to 23 mm Hg six months later (table 1). The pulmonary valve showed features of commissural fusion in addition to dysplasia. On the angiogram the base of one of the leaflets appeared to be thin and a systolic jet was noted (fig 1). An echocardiogram (fig 1) of the pulmonary valve from the long axis view showed doming as well as leaflet thickening, with widening of one of the sinuses of Valsalva in diastole. Thus although the valve leaflets were thickened, there was an element of commissural fusion to account for the success of balloon dilatation.

Table 2 shows the angiographic and echocardiographic features in these patients. All patients showed angiographic evidence of thickening of pulmonary valve leaflets. A supra-annular ridge was noted in all but one patient (case 7) (fig 2). Patients 5 and 7 did not show asymmetric systolic doming, and a systolic jet was seen in four of the seven patients. Post-stenotic dilatation of the proximal main pulmonary artery was present in two patients (cases 4 and 7). Patients 5 and 7 showed diastolic widening of one sinus of Valsalva on the lateral projection of the right ventriculogram, suggesting that in addition to dysplasia there was commissural fusion between at least two adjacent cusps. None of the patients had fixed subvalve pulmonary stenosis. The mean (SD) diameter of the pulmonary valve annulus was 10·7 (2·11) mm and this is 90 (18) % of the predicted normal for body weight.[19]

Table 2 *A comparison of angiographic and echocardiographic features in seven patients with dysplastic pulmonary valves*

	Patient No:						
	1	*2*	*3*	*4*	*5*	*6*	*7*
Angiographic features:							
Thickened leaflets	+	+	+	+	+	+	+
Asymmetrical systolic doming	+	+	+	+	○	+	○
Supra-annular narrowing	+	+	+	+	+	+	○
Systolic jet	○	○	+	+	+	○	+
Post-stenotic dilatation	○	○	○	+	○	○	+
Echocardiographic features:							
Thickened leaflets	+	+	+	+	+	+	+
Immobility of leaflets	+	+	+	+	○	+	○
Supra-annular narrowing	+	+	+	+	+	+	○
Post-stenotic dilatation	○	○	○	○	○	○	+

+, present; ○, absent.

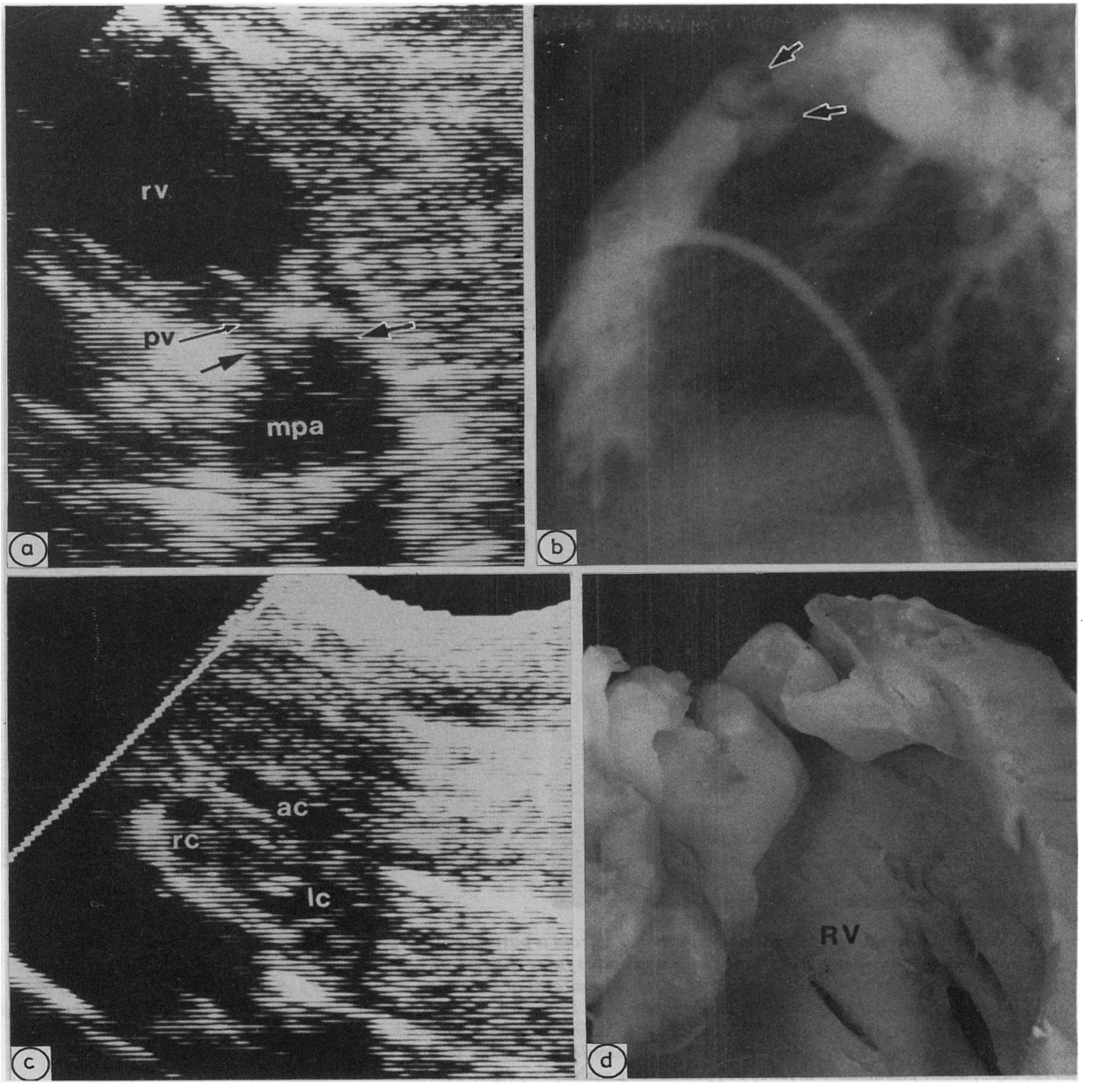

Fig 2 *(a) Echocardiogram of precordial long axis view from a patient with a dysplastic pulmonary valve. Note the bright echoes originating from the thickened valve. There is also evidence of supravalvar narrowing (black arrows). (b) Angiocardiogram in the same patient. Note the absence of a central jet, the supravalvar narrowing (black arrows), and a fleshy appearance of the valve leaflets. (c) Short axis echocardiographic view. (d) Photograph of long axis cut from another patient with a dysplastic pulmonary valve showing fleshy and thickened valve leaflets. There is no evidence of fusion along the commissures. ac, anterior cusp; lc, left cusp; rc, right cusp; mpa, main pulmonary artery; pv, pulmonary valve; rv, right ventricle.*

The cross sectional echocardiographic features that were common to patients with dysplasia correlated closely with those seen during ventriculography (figs 1–2 and table 2). The high parasternal short axis view revealed tricuspid pulmonary valves in all patients. Both the shape and the mobility of individual valve leaflets could be determined by combining the parasternal long and short axis views;

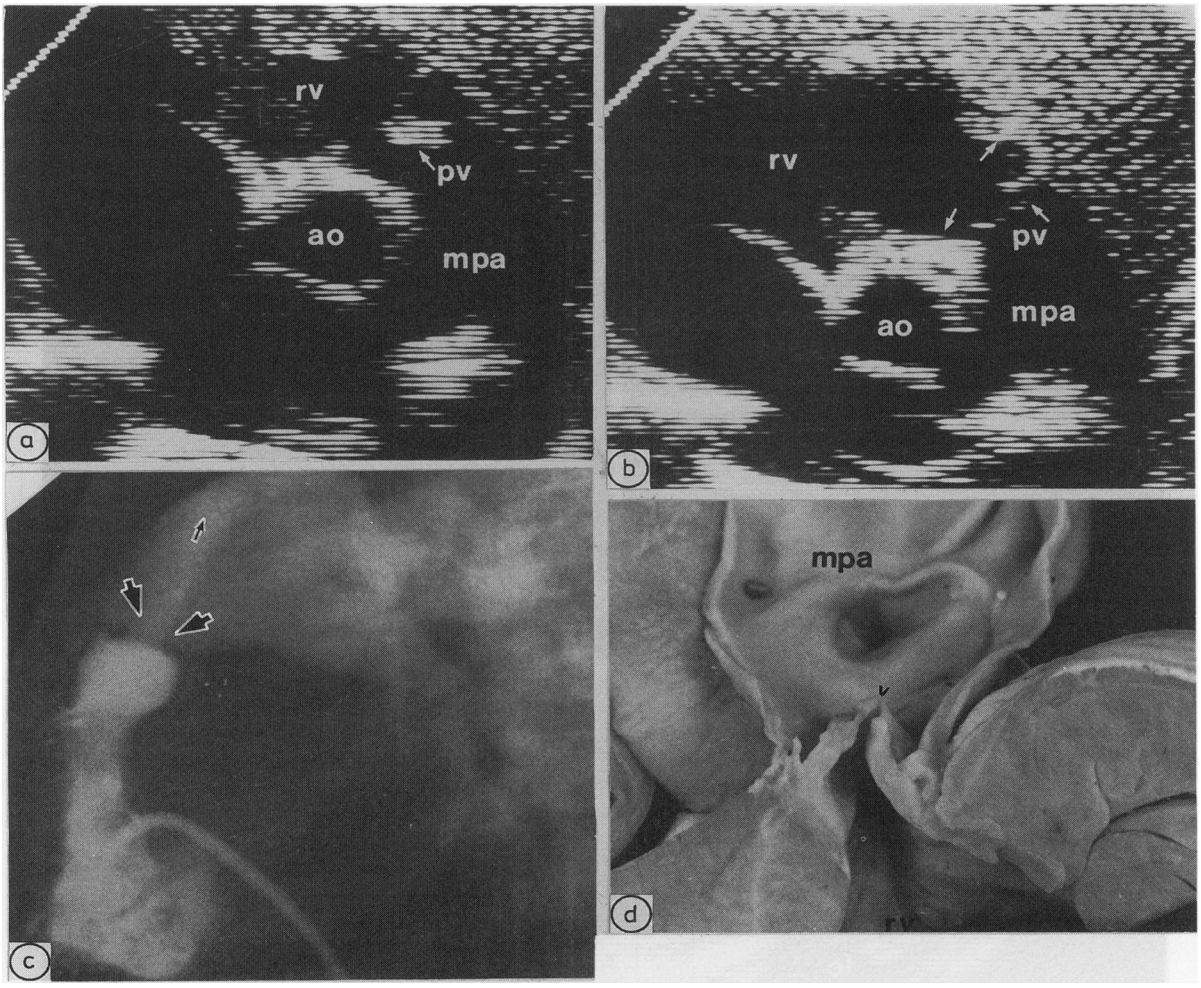

Fig 3 *(a) Echocardiogram of precordial short axis view from a patient with typical pulmonary valve stenosis. Note the bright echoes at the tips of the leaflets where the commissures fuse; the remainder of the leaflet appears to be thin. (b) Valve doming in the same patient; the white arrows indicate the margins of the valve annulus. (c) Angiocardiogram showing the central jet typical of isolated pulmonary valve stenosis. (d) Photograph of a specimen cut in the long axis from a neonate with critical pulmonary valve stenosis. Note the pinpoint orifice indicated by the black arrow. The valve leaflets are slightly thickened; however, they are very different from the fleshy leaflets in the patient with a dysplastic valve seen in fig 2. ao, aorta; mpa, main pulmonary artery, pv, pulmonary valve; rv, right ventricle.*

all patients had thickened leaflets. Poor leaflet mobility could be seen in both long and short axis views in all patients except cases 5 and 7. The anterior sinus of Valsalva in each of these patients showed diastolic widening in the parasternal long axis view—a feature not seen in either the posterior sinuses of Valsalva in these two cases or in any of the other patients. Post-stenotic dilatation of the main pulmonary artery was seen in one patient only (case 7).

Discussion

The association of a particular phenotype with pulmonary valve stenosis has been well recognised since the original description by Noonan in the early 1960s, with up to 50% of patients having dysplasia of the valve as the mechanism of stenosis.[7 8] It is very likely that the wide variation seen in the physical features of this syndrome is similarly expressed in variable degrees of valve dysplasia.[3] Other clinical

features distinguishing dysplasia from typical (commissural fusion) pulmonary valve stenosis are absence of an ejection click, a frontal QRS axis of $>170°$ on surface electrocardiogram, and lack of post-stenotic dilatation of the main pulmonary artery on the chest radiograph.[4-6 9]

The gold standard of diagnosis, however, remains the angiographic demonstration of considerably thickened valve leaflets, irregular asymmetrical doming in systole, absence of diastolic widening of sinuses of Valsalva, and a hypoplastic valve annulus and proximal main pulmonary artery.[5] Furthermore, a systolic contrast jet is occasionally seen. On the basis of necropsy examinations,[4] the mechanism of obstruction is thought to result from the thickening of the leaflets, which are confined within a hypoplastic valve annulus, with little or no commissural fusion.[4 6 9] The echocardiographic features for the diagnosis of pulmonary valve dysplasia correlate well with the angiographic features. They are pronounced leaflet thickening combined with lack of diastolic widening of the sinuses of Valsalva and supra-annular narrowing. Poor mobility of pulmonary valve leaflets was seen in both the parasternal long and high short axis views. Evidence of diastolic widening of one or more of the sinuses of Valsalva in the long axis view suggests that commissural fusion, as well as a leaflet abnormality, may be a part of the mechanism of obstruction. These observations are in direct contrast with those seen in typical isolated pulmonary valve stenosis in which the mechanism of stenosis is commissural fusion (fig 3).

The application of balloon dilatation to dysplasia of the pulmonary valve was disappointing in our series and in the five patients reported by other investigators.[14 15 17] This is not surprising because disruption of commissural fusion appears to be the mechanism for relief of obstruction in this condition.[18] Valve annulus hypoplasia cannot account for residual or persistent stenosis because hypoplasia (that is <2SD below expected normal) was not a prominent feature in our patients, although this feature was noted by others.[2-6] Furthermore, among patients with pulmonary valve stenosis, there will be a number with mixed dysplasia and commissural fusion. It is of interest that 20% of patients with valve dysplasia described by Jeffrey *et al*, showed angiographic systolic jetting,[5] a feature found in $>90\%$ of cases of typical doming pulmonary valve stenosis. Similarly Schneeweiss *et al* discussed 12 anatomically confirmed cases of pulmonary valve dysplasia and showed post-stenotic dilatation in 75% and systolic jetting in 33%.[20] The traditional angiographic features of pulmonary valve dysplasia may therefore reflect some contribution by commissural fusion.

Patients with mixed commissural fusion and dysplasia may obtain relief of stenosis from balloon dilatation of the valve. Cross sectional echocardiographic imaging of the pulmonary valve, particularly in the parasternal long axis view, allows adequate imaging of at least two of the leaflets (figs 1 and 3). Where echocardiographic imaging demonstrates features suggestive of dysplasia, diastolic frames should be carefully examined for widening of the sinuses of Valsalva. This feature we believe indicates that commissural fusion is part of the mechanism of obstruction. In those instances, cardiac catheterisation and angiography should be undertaken to confirm the presence of commissural fusion and dysplasia and balloon dilatation should be considered when gradient reduction is regarded as clinically appropriate.

References

1 Campbell M. Simple pulmonary stenosis. Pulmonary vascular stenosis with a closed ventricular septum. *Br Heart J* 1954;**16**:273–300.

2 Noonan JA. Hypertelorism with Turner phenotype. *Am J Dis Child* 1968;**116**:373–80.

3 Linde LM, Turner SW, Sparkes RS. Pulmonary valvular dysplasia. A cardiofacial syndrome. *Br Heart J* 1973;**35**:301–4.

4 Koretzky ED, Moller JH, Korns ME, Schwartz CJ, Edwards JE. Congenital pulmonary stenosis resulting from dysplasia of the valve. *Circulation* 1969; **40**:43–53.

5 Jeffrey RF, Moller JH, Amplatz K. The dysplastic pulmonary valve: a new roentgenographic entity. *Am J Roentgenol Rad Therapy Nuclear Med* 1972;**114**: 322–39.

6 Schieken RM, Friedman S, Pierce WS. Severe congenital pulmonary stenosis with pulmonary valvular dysplasia syndrome. *Ann Thorac Surg* 1973;**15**: 570–7.

7 Duncan WJ, Fowler RS, Farkas LG, *et al*. A comprehensive scoring system for evaluating Noonan syndrome. *Am J Med Genet* 1981;**10**:37–50.

8 Nora JJ, Nora AH, Sinha AK, Spangles RD, Lubs HA. The Ullrich-Noonan syndrome (Turner phenotype). *Am J Dis Child* 1974;**127**:48–55.

9 Watkins L Jr, Donahoo JS, Harrington D, Haller JA, Neill CA. Surgical management of congenital pulmonary valve dysplasia. *Ann Thorac Surg* 1977; **21**:498–507.

10 Mistrot J, Neal W, Lyons G, *et al*. Pulmonary valvulotomy under inflow stasis for isolated pulmonary stenosis. *Ann Thorac Surg* 1976;**21**:30–7.

11 Vancini M, Roberts KD, Silove ED, Singh SP. Surgical treatment of congenital pulmonary stenosis due to dysplastic leaflets and small valve annulus. *J Thorac Cardiovasc Surg* 1980;**79**:464–8.

12 Kan JS, White RI Jr, Mitchell SE, Gardner TJ.

Percutaneous balloon valvuloplasty: a new method for treating congenital pulmonary valve stenosis. *N Engl J Med* 1982;**307**:540–2.

13 Labadidi Z, Jiunn-Ren W. Percutaneous balloon pulmonary valuloplasty. *Am J Cardiol* 1983;**52**:560–2.

14 Kan JS, White RI Jr, Mitchell SE, Anderson JH, Gardner TJ. Percutaneous transluminal balloon valvuloplasty for pulmonary valve stenosis. *Circulation* 1984;**69**:554–60.

15 Walls JT, Labadidi Z, Curtis JJ, Silver D. Assessment of percutaneous balloon pulmonary and aortic valvuloplasty. *J Thorac Cardiovasc Surg* 1984;**88**:352–6.

16 Rocchini AP, Kveselis DA, Crowley D, Dick M, Rosenthal A. Percutaneous balloon valvuloplasty for treatment of congenital pulmonary valvular stenosis in children. *J Am Coll Cardiol* 1984;**3**:1005–12.

17 Tynan M, Baker EJ, Rohmer J, *et al*. Percutaneous balloon pulmonary valvuloplasty. *Br Heart J* 1985; **53**:520–4.

18 Benson LN, Smallhorn JS, Freedom RM, Trusler GA, Rowe RD. Pulmonary valve morphology after balloon dilatation of pulmonary valve stenosis. *Cathet Cardiovasc Diagn* 1985;**11**:161–6.

19 Rowlatt VF, Rimoldi HJA, Lev M. The quantitative anatomy of the normal child's heart. *Pediatr Clin N Am* 1963;**10**:499.

20 Schneeweiss A, Blisden LC, Shem-Tov A, Orr D, Milo S, Neufeld HN. Diagnostic angiocardiographic criteria in dysplastic pulmonic valve. *Am Heart J* 1985;**106**:761–2.